

FLAER

Paroxysmal
Nocturnal
Hemoglobinuria

- **Alexa fluor® 488 labelled reagent**
- **More sensitive than CD59;
detects small, abnormal granulocyte
populations to a level of 0.5%**
- **Now available in stabilized
liquid and lyophilized formats**

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ISO 9001 and ISO 13485 registered.

PNH: A progressive, destructive, and life-threatening disease

Paroxysmal nocturnal hemoglobinuria (PNH) is a stem cell disorder caused by a mutation of a gene involved in the synthesis of the GPI (glycosylphosphatidylinositol) anchor of a group of surface proteins on circulating cells. Affected cells are sensitive to complement-mediated hemolysis and this may lead to life-threatening thrombosis, chronic kidney disease, pulmonary hypertension, end organ damage, ischemic bowel disease, hepatic failure, and anemia—all of which contribute to a shortened lifespan for patients with PNH.

Identifying PNH patients early in the course of their disease may offer the best opportunity for long-term management. In the past, PNH has been challenging to identify effectively. However, in recent years impressive strides have been made in understanding of PNH pathology, accompanied by greatly improved detection techniques, including multiparametric flow cytometry using FLAER.

About FLAER

FLAER is an Alexa® 488 labeled variant of aerolysin, a unique protein that binds tightly and specifically to mammalian GPI anchors. FLAER will not bind to PNH cells because they do not produce the anchor. Before FLAER, detection of PNH clones by flow cytometry relied on fluorescently labeled antibodies to GPI-linked proteins such as CD59 and CD55. These antibodies do not bind with high affinity, so that small PNH clones are not detected. Also, they each screen for the absence of a specific protein, rather than loss of the GPI anchor, and therefore there is the risk of false negative results. Since FLAER binds to the GPI anchor itself, only PNH cells, which lack the anchor, will be negative. And since FLAER binds with high affinity, very small PNH populations can be detected.

Liquid FLAER and reconstituted improved powdered FLAER are equivalent. Both retain strong signals after aAt least 4 months when storAed at 4°C.

FLAER (Alexa 488 proaerolysin variant)

Format	Size	Est. # of Tests	Cat #
Powder	25 ug	50	CL-FL1
Powder	50 ug	100	CL-FL2
Liquid	25 ug	50	CL-FL1S
Liquid	50 ug	100	CL-FL2S

The products are for Research Use Only

References

A recent brief review of the status and importance of PNH and the key role of FLAER in detection by flow cytometry:

Titus, K. (2009) *Bringing wider meaning to a rare disease. Cap Today. January*

A recent published method for PNH detection using FLAER:

Sutherland et al. (2007) *Diagnosing PNH with FLAER and multiparameter flow cytometry. Cytometry Part B (Clinical Cytometry) 72B: 167-177*

A detailed protocol is available upon request.

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Camino del Pilón, 86 Casa- 7

50011- Zaragoza

e-mail: dlw@dlongwood.com

Tel. 976 320 638 Fax. 976 320 660